Streptococcal Toxic-Shock Syndrome
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Overview[1,2]

For a complete description of Streptococcal Toxic-Shock Syndrome, refer to the following text:

- Control of Communicable Disease Manual (CCDM).
- Red Book, Report of the Committee on Infectious Diseases.

Case Definition [3]

Clinical description [1]

Streptococcal toxic-shock syndrome (STSS) is a severe illness associated with invasive or noninvasive group A streptococcal (*Streptococcus pyogenes*) infection. STSS may occur with infection at any site but most often occurs in association with infection of a cutaneous lesion. Signs of toxicity and a rapidly progressive clinical course are characteristic, and the case-fatality rate may exceed 50%.

Clinical case definition

An illness with the following clinical manifestations occurring within the first 48 hours of hospitalization or, for a nosocomial case, within the first 48 hours of illness:

- Hypotension defined by a systolic blood pressure £90 mm Hg for adults or less than the fifth percentile by age for children aged <16 years Multi-organ involvement characterized by two or more of the following:
    1. Renal impairment: Creatinine ≥2 mg/dL (≥177 μmol/L) for adults or greater than or equal to twice the upper limit of normal for age. In patients with preexisting renal disease, a greater than twofold elevation over the baseline level.
    2. Coagulopathy: Platelets ≤100,000/mm^3 (≤100 x 10^6 /L) or disseminated intravascular coagulation, defined by prolonged clotting times, low fibrinogen level, and the presence of fibrin degradation products
    3. Liver involvement: Alanine aminotransferase, aspartate aminotransferase, or total bilirubin levels greater than or equal to twice the upper limit of normal for the patient's age. In patients with preexisting liver disease, a greater than two-fold increase over the baseline level
    4. Acute respiratory distress syndrome: defined by acute onset of diffuse pulmonary infiltrates and hypoxemia in the absence of cardiac failure or by evidence of diffuse capillary leak manifested by acute onset of generalized edema, or pleural or peritoneal effusions with hypoalbuminemia
    5. A generalized erythematous macular rash that may desquamate
    6. Soft-tissue necrosis, including necrotizing fasciitis or myositis, or gangrene

Case classification
Probable: a case that meets the clinical case definition in the absence of another identified etiology for the illness and with isolation of group A *Streptococcus* from a nonsterile site
Confirmed: a case that meets the clinical case definition and with isolation of group A *Streptococcus* from a normally sterile site (e.g., blood or cerebrospinal fluid or, less commonly, joint, pleural, or pericardial fluid) [1]

Laboratory criteria for diagnosis

Isolation of group A *Streptococcus*

Discussion

Most cases of STSS arise from an initial soft tissue infection. Most cases occur in previously healthy individuals. Puerperal infections, in some cases requiring hysterectomy or progressing to fatal disseminated coagulation are also documented. Most cases occur in adults, although cases have been reported in children. Even in children, cases are usually the result of soft tissue infections, especially secondary infection of chicken pox lesions. [4, 5]

STSS is most often caused by the M1 serotype of *Streptococcus pyogenes*, although other serotypes and even (rarely) strains of group B or group C strep have been implicated. As a result, unlike Staphylococcal Toxic Shock Syndrome, STSS can be transmitted from person to person. Clusters within families or in the nosocomial setting, such as in nursing homes, have been reported. In one instance, an emergency medical technician was infected after attempting to resuscitate of a child with the syndrome. [4]

The progress of this syndrome is extremely rapid and can lead rapidly to death. In addition, because many cases of STSS occur in patients with invasive soft tissue infections, the clinical features of STSS and necrotizing fasciitis often overlap. Typically, the infection begins at the site of a minor, even non-penetrating trauma or as the result of pneumonia. A prodromal phase of influenza-like illness is reported in 20% of patients.

Within 24 – 72 hours, the case presents with intense pain, high fever, prostration, localized swelling and erythema. Mandell[4] says that the development of vesicles and the appearance of a violaceous or bluish hue are ominous findings. Concurrent or shortly after the initial presentation, hypotension and numerous signs and symptoms of multiorgan failure appear. Without rapid and aggressive hospital treatment, the case fatality rate is very high.

Reporting Requirements

Report as with any invasive Group A Streptococcal infection. This is a category I disease and must be reported to the local health authority or the Missouri Department of Health within 24 hours of first knowledge or suspicion by telephone, facsimile, or other rapid communication.

1. If the case develops STSS,contact your District Communicable Disease Coordinator immediately.

2. For confirmed and probable cases, complete a "MDOH Disease Case Report" form (CD-1).
3. For each case, gather information needed to complete a "Record of Investigation of Communicable Disease" form (CD-2).
4. Send completed forms to the District Health Office.
5. Entry of appropriate information into the MOHSIS database will satisfy items 1, 2, and 3 of this section.
6. Within 90 days from the conclusion of an outbreak, submit the final outbreak report to the District Communicable Disease Coordinator.

References

1. Centers for Disease Control. Case Definitions for Infectious Conditions Under Public Health Surveillance. MMWR 1997; 33-34 (RR-10)

Group A Streptococcal (GAS) Disease

Fact Sheet

What is group A streptococcus (GAS)?

Group A streptococci are bacteria often found in the throat and on the skin. People may carry group A streptococci in the throat or on the skin and have no symptoms of disease. The vast majority of GAS infections are relatively mild illnesses, such as strep throat and impetigo. On rare occasions, these bacteria can cause much more severe and even life-threatening diseases such as necrotizing fasciitis or streptococcal toxic shock syndrome (STSS).

How are group A streptococci spread?

These bacteria are spread through direct contact with mucus from the nose or throat of persons who are infected or through contact with infected wounds or sores on the skin. Ill persons, such as those who have strep throat or skin infections, are most likely to spread the infection. Persons who carry the bacteria but have no symptoms are much less contagious. Treating an infected person with an antibiotic for 24 hours or longer generally eliminates their ability to spread the bacteria. However, it is important to complete the entire course of antibiotics as prescribed. It is not likely that household items like plates, cups, or toys spread these bacteria.

What kind of illnesses are caused by group A streptococcal infection?

Infection with GAS can result in a range of symptoms:
No illness
Mild illness (strep throat or a skin infection such as impetigo)
Severe illness (necrotizing faciitis, streptococcal toxic shock syndrome)

Severe, sometimes life-threatening, GAS disease may occur when bacteria get into parts of the body where bacteria usually are not found, such as the blood, muscle, or the lungs. These infections are termed "invasive GAS disease." Two of the most severe, but least common, forms of invasive GAS disease are necrotizing fasciitis and STSS. Necrotizing fasciitis (occasionally described by the media as "the flesh-eating bacteria") destroys muscles, fat, and skin tissue. STSS causes blood pressure to drop rapidly and organs (e.g., kidney, liver, lungs) to fail. STSS is not the same as the "toxic shock syndrome" frequently associated with tampon usage. About 20% of patients with necrotizing fasciitis and more than half with STSS die. About 10%-15% of patients with other forms of invasive group A streptococcal disease die.

How common is invasive group A streptococcal disease?

About 10,000 cases of invasive GAS disease occurred in the United States in 1998. Of these, about 600 were STSS and 800 were necrotizing fasciitis. In contrast, there are several million cases of strep throat and impetigo each year. In 1998 there were 18 cases of invasive GAS disease reported in Missouri.

Why does invasive group A streptococcal disease occur?

Invasive GAS infections occur when the bacteria get past the defenses of the person who is infected. This may occur when a person has sores or other breaks in the skin that allow the bacteria to get into the tissue, or when the person’s ability to fight off the infection is decreased because of chronic illness or an illness that affects the immune system. Also, some virulent strains of GAS are more likely to cause severe disease than others.

Who is most at risk of invasive group A streptococcal disease?

Few people who come in contact with a virulent strain of GAS will develop invasive GAS disease; most will have a routine throat or skin infection and some may have no symptoms whatsoever. Although healthy people can get invasive GAS disease, people with chronic illnesses like cancer, and diabetes, people receiving kidney dialysis, and those who use medications such as steroids, are at higher risk. In addition, breaks in the skin, like cuts, surgical wounds or chickenpox may provide an opportunity for the bacteria to enter the body.

What are the early signs and symptoms of necrotizing fasciitis and streptococcal toxic shock syndrome?

Early signs and symptoms of necrotizing fasciitis:
Fever
Severe pain and swelling
Redness at a wound site

Early signs and symptoms of STSS:
Fever
Dizziness
Confusion
A flat red rash over large areas of the body

How is invasive group A streptococcal disease treated?

GAS infections can be treated with many different antibiotics. Early treatment may reduce the risk of death from invasive group A streptococcal disease. However, even the best medical care does not prevent death in every case. For those with very severe illness, supportive care in an intensive care unit may be needed. For persons with necrotizing fasciitis, surgery often is needed to remove damaged tissue.

What can be done to help prevent group A streptococcal infections?

The spread of all types of GAS infection can be reduced by good hand washing, especially after coughing and sneezing and before preparing foods or eating. Persons with sore throats should be seen by a doctor who can perform tests to find out whether the illness is strep throat. If the test result shows strep throat, the person should stay home from work, school, or child care until 24 hours after taking an antibiotic. All wounds should be kept clean and watched for possible signs of infection such as redness, swelling, drainage, and pain at the wound site. A person with signs of an infected wound, especially if fever occurs, should seek medical care. It is not necessary for all persons exposed to someone with an invasive group A strep infection (i.e. necrotizing fasciitis or strep toxic shock syndrome) to receive antibiotic therapy to prevent infection. However, in certain circumstances, antibiotic therapy may be appropriate. That decision should be made after consulting with your doctor.

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